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TROPHONEUROTIC CHANGES IN BONES AND JOINTS IN LEPROSY

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TROPHONEUROTIC CHANGES IN BONES AND JOINTS IN LEPROSY *

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It is well recognized that in leprosy, especially its anesthetic form, marked deformities of the hands and feet may develop. Such cases are generally designated as "lepra mutilans." Many, perhaps the majority of these cases, are dependent on ulceration, necrosis, and chronic suppurative inflammation, which in turn are indirectly due to a chronic leprous neuritis with its accompanying anesthesia and analgesia. Necroses or ulcers form, phalanges gradually slough off, and finally the characteristic, clubbed hands and feet of leprosy are formed. The accompanying illustrations of such hands and feet show their appearance in some very pronounced cases. This explanation does not hold in all cases, however, as some of these deformities are to be ascribed to a peculiar trophoneurotic influence which is probably directly exerted by the neuritis on the tissues, especially those of the osseous system, and which may result in atrophy of bone of characteristic appearance, and in certain joint affections.

The first to call attention to the occurrence of such atrophies of bone in leprosy was the late Prof. H. Heiberg,[1] whose work on this subject, published in 1886, has received little attention, although it contains interesting observations and touches on questions of importance in physiology and general pathology. Since then, little has been written on this subject (Deycke, Hirschberg and Bichler). I therefore have desired to renew the study of this problem[2] and questions arising with it, and will present the results in the following pages. I feel especially inclined to do this, having had at my disposal most excellent, and in some respects, unique material, partly from our pathologic institute in Christiania, and partly from the leprosy hospital in Bergen, which latter material was

* An abstract of this paper, illustrated by specimens, photographs and diapositives, was read before the Second International Leprosy Conference, held at Bergen, Norway, Aug. 16-19, 1909.

1. Heiberg, H.: Om lepra mutilans og trofoneurotiske forandringer ved spedalskhed, Klin. Aarborg., 1886, iii.

2. I have discussed this subject once before, in my competitive thesis for the professorship in pathologic anatomy and general pathology, entitled Om de pathologisk-anatomiske forandringer af neurotrofisk oprindelse, Christiania. 1900, A. W. Brögger's Buchdruckerei. This work is especially devoted to microscopic investigations of these atrophies.

kindly placed at my disposal by Dr. H. P. Lie. As will be seen by the case reports, the material was obtained from patients who had suffered from leprosy from fifteen to sixty-seven years, during most of which time they had been under observation in our leprosy hospitals.

SPECIMENS

Specimen 1.—Concentric atrophy of finger phalanges (Fig. 1). The skeleton of the right hand of a leprous patient. The carpus is on the whole normal. The metacarpal bones also have their usual appearance, but are somewhat thin, rather short, especially in the diaphyses, but have capitula of usual appearance. They measure in length (from No. 1 to 5, respectively) 4.5 cm., 6.4 cm., 6 cm., 5.8 cm. and 5.2 cm. Of the phalanges, only the first one is preserved and merely

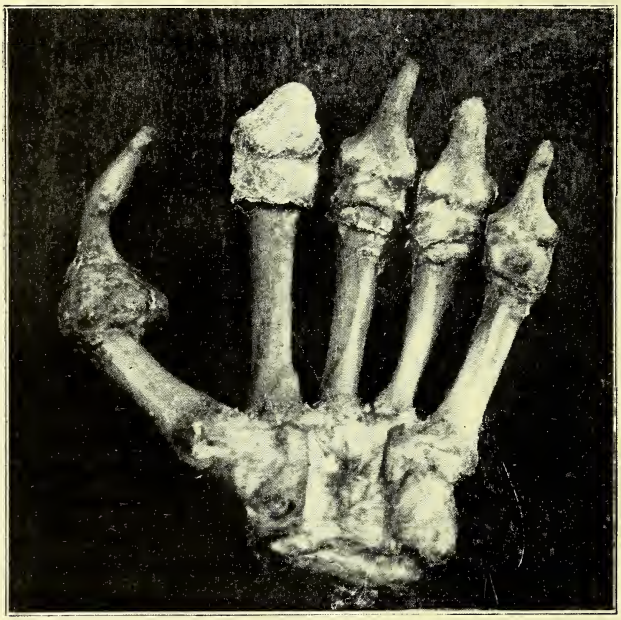

Fig. 1.—Right hand; concentric atrophy of the phalanges.

the shape of pieces of bone markedly pointed anteriorly, and measuring 3.8 cm. (phalanges grown together), 1.4 cm., 2.5 cm., 2.2 cm. and 2 cm. Hence the second and third phalanges are missing in the last four fingers.

Specimen 2.—Atrophy of the finger phalanges (Fig. 2). Left hand of a patient with anesthetic leprosy who contracted the disease in 1820, was admitted to the Reknäs Hospital for lepers in 1875, and died of peritonitis in 1887, sixty-seven years after the onset of leprosy. The feet were greatly deformed on account of extensive carious processes. The left hand was club-shaped on account of necrosis of the phalanges. The specimen shows that the carpus is preserved, and the metacarpal bones are about normal. The fingers are flexed at right angles at the metacarpophalangeal joints, pointed anteriorly, and defective. The measurements are as follows:

Os metacarpi I, 5 cm.; Phalanx I, 3.6 cm.; Phalanx II, 2 cm. (normal).

Os metacarpi II, 7.2 cm.; only one phalanx, 3 cm. long, markedly pointed anteriorly.

Os metacarpi III, 6.8 cm.; only one rudiment of a phalanx.

Os metacarpi IV, 6 cm.; only one phalanx (No. 1 ?), 4.6 cm. long, of usual appearance.

Os metacarpi V, 5.2 cm.; only one phalanx (No. 1 ?), 3.5 cm. long, of usual form with a small rudiment anteriorly 1.4 cm. long.

Specimen 3.—Concentric atrophy of the phalanges of the fingers (Fig. 3). Right hand from a patient with lepra anesthetica (patient whose foot is described as Specimen 8). The hand is deformed, shortened, extended at the wrist, and

Fig. 2.—Left hand; atrophy of phalanges.

the fingers are greatly shortened. The otherwise normal carpus, here and there, presents knotty projections (in part due to chronic arthritis deformans?). The metacarpal bones are nearly normal, but the phalanges are few in number, small, pointed anteriorly, and partly placed at right angles to the metacarpal bones. The measurements are as follows:

Os metacarpi I, 4.5 cm. long; both phalanges missing.

Os metacarpi II, 6.1 cm. long; only one phalanx, 1.8 cm. long, pointed anteriorly.

Os metacarpi III, 5 cm. long; one phalanx (No. 1 ?), 1.8 cm. long.
Os metacarpi IV, 5.6 cm. long; one phalanx 2.1 cm. long, pointed anteriorly.
Os metacarpi V, 5 cm. long; one phalanx 2 cm. long.

Specimen 4.—Some degree of atrophy of the finger phalanges (Fig. 4). The specimen is the left hand (the right foot is Specimen 5) of a patient with lepra anesthetica with onset in 1861, admittance to a leprous hospital in 1864, and death in 1887, or twenty-six years after the onset. Already in 1864, there was considerable paresis of the face, the upper and particularly the lower extremities, with loss of sensation. The left hand, removed at necropsy, shows the following conditions: The carpus is entirely normal. The metacarpal and phalangeal bones also are of about usual size and shape. The position, with the fingers markedly flexed, is that known as "claw-hand."

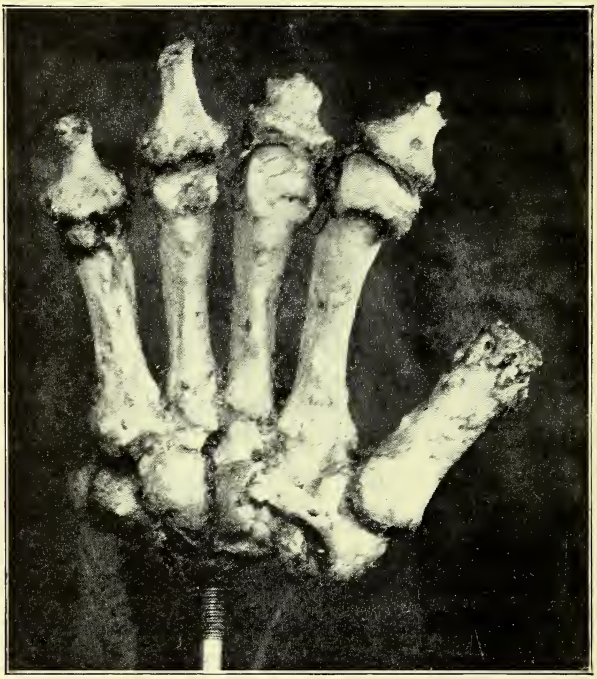

Fig. 3.—Right hand; concentric atrophy of phalanges.

Os metacarpi I, 4.6 cm.; Phalanx I, 3 cm.; Phalanx II, 2 cm.
Os metacarpi II, 6.3 cm.; Phalanx I, 2 cm., pointed; Phalanx II, 1.5 cm., thin in the middle; Phalanx III, 1.4 cm., pointed.
Os metacarpi III, 6.2 cm.; Phalanx I, 4.7 cm., pointed; Phalanx II, 3.1 cm.; Phalanx III, 1.6 cm.
Os metacarpi IV, 5.3 cm.; Phalanx I, 3.2 cm.; Phalanx II, 2.6 cm.; Phalanx III, 1.7 cm.; all bones nearly normal.
Os metacarpi V, 5 cm.; Phalanx I, 3.5 cm.; Phalanx II, 2.4 cm.; Phalanx III, 1.2 cm.; all bones nearly normal.

Specimen 5.—Ulcerative-necrotic inflammation of the tarsus (lepra mutilans); typical concentric atrophy of the fourth and fifth metatarsal bones and loss of phalanges of the fifth toe. In the right foot (same patient as Specimen 4), there

had been a carious-necrotic process, so that the foot at necropsy was found to be loosened at the ankle, being attached only by the soft parts. The foot was of about normal length, but unusually narrow on account of atrophy of its outer portion. The tarsus shows great changes. The calcaneum is 7.8 cm. long, tapers forward, and shows necrosis on the inner side and at the anterior end. The talus, os cuboideum, and os naviculare, as well as the cuneiform bones II and III, are nearly entirely wanting, while a considerable part of os cuneiforme I is intact. The greater part of the metatarsal bones is preserved; there is atrophy only on the fibular side.

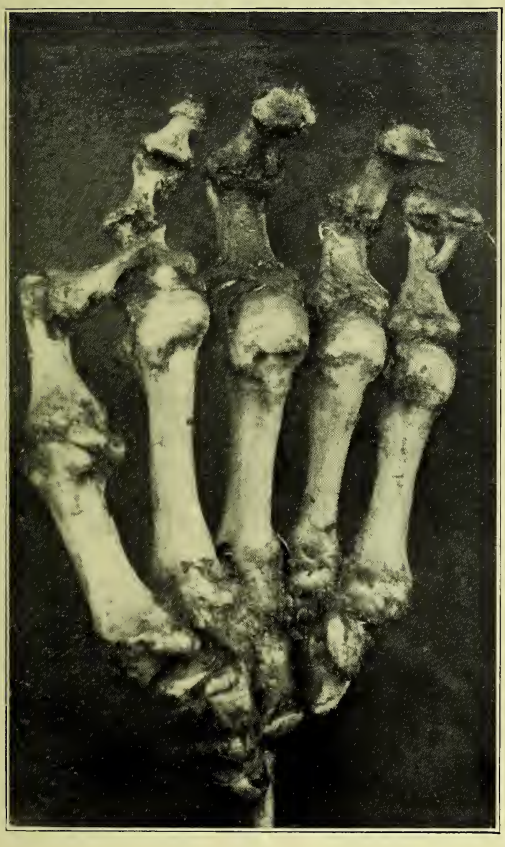

Figure 4.

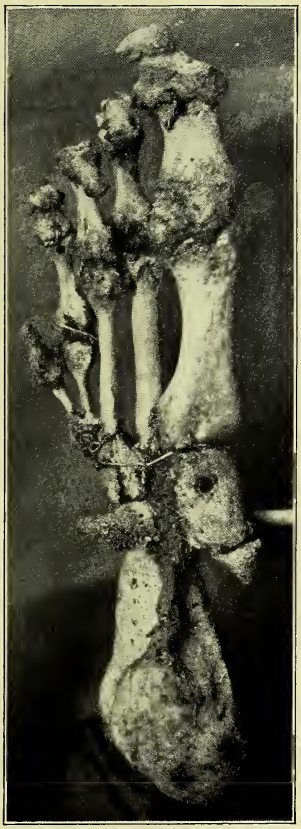

Figure 5.

Fig. 4.—Left hand; moderate atrophy of phalanges.
Fig. 5.—Right foot; lepra mutilans; concentric atrophy of fourth and fifth metatarsal bones.

Os metacarpi I, 6.2 cm.; Phalanx I, 3.3; Phalanx II, 2.5 cm.; all nearly normal.

Os metacarpi II, 7 cm.; Phalanx I, 3.2 cm.; Phalanx II, 1.8 cm.; Phalanx III, 1.2 cm.; all nearly normal.

Os metacarpi III, 6.8 cm.; Phalanx I, 2.7; Phalanx II, 1.6 cm.; Phalanx III, 1.4 cm.; all nearly normal.

Os metacarpi IV, 3.2 cm.; small, atrophic, with thin shafts like narrow edges; knobbed anterior ends; Phalanx I, 2.6 cm.; Phalanx II, 1.5 cm.; Phalanx III, 1.2 cm.

Os metacarpi V, 2.5 cm.; small, atrophic, with thin shafts like narrow edges; knobbed anterior ends; Phalanx I, 1.7, atrophic and pointed anteriorly; both the other phalanges missing.

Figure 6.

Figure 7.

Fig. 6.—Left foot; concentric peripheral atrophy of metatarsal bones and phalanges.

Fig. 7.—Right foot; atrophy of metatarsal bones and phalanges.

Specimen 6.—Previously described in brief by H. Heiberg. Typical concentric peripheral atrophy of the metatarsal bones and of the phalanges (Fig. 6). The skeleton of the left foot of a leprous man 38 years old. The foot is small,

measuring 17 cm. in length. There is concentric atrophy of the metatarsal bones and phalanges, especially in their anterior portions. The entire tarsus shows normal conditions with well-developed bones and joints. The tarsal bones on the whole are of normal size, although the calcaneum is possibly somewhat short. The metatarsal bones, on the other hand, are small, atrophic, and greatly

Figure 8.

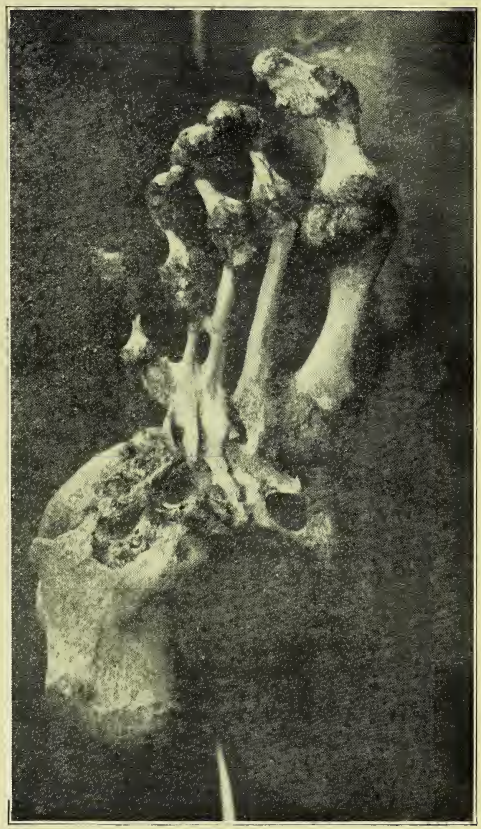

Figure 9.

Fig. 8.—Left foot; concentric atrophy of metarsal bones and phalanges.
Fig. 9.—Right foot; ulcerative-necrotic inflammation of tarsus; atrophy of several metarsal bones.

shortened. They measure, respectively, 2.8 cm., 3.7 cm., 3.6 cm., 4.4 cm., 6.3 cm., and compared to the average length of the metatarsal bones in four normal feet, the shortening is, respectively, 3.5 cm., 3.9 cm., 2.6 cm., 1.8 cm. Their shape is also considerably changed. Their width at the base is about normal, but they taper

considerably and unevenly forward and become pointed in a conical manner. The heads of the metatarsal bones are nearly entirely absent, as far as the three middle toes are concerned. The surface of the bones as a rule is smooth. Only rudiments of the phalanges are left, which are atrophic, thin and pointed, tapering forward. In places they are so thin that a thin wire can scarcely be passed through them. The osseous substance is porous. Only small remnants remain of some of the bones. If the atrophying process had continued, the bones certainly would have become entirely resorbed.

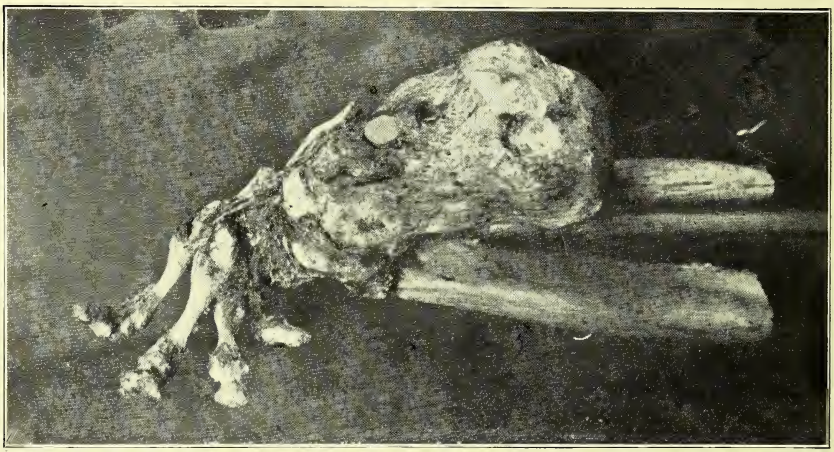

Fig. 10.—Left foot; lepra mutilans.

Fig. 11.—Left foot; same as Fig. 10, showing fusion of tibia and fibula.

Specimen 7.—Typical atrophy of metatarsal bones and phalanges. Right foot with tendons and joints from a leprous patient. The foot is small and atrophic, partly on account of defective and hyperextended toes, and partly from a considerable atrophy of the metatarsal bones. The tarsus on the whole is normal; the bones are well developed without nodular projections. The various joints show no actual signs of arthritis. The tendons and ligaments are normal. On the other hand, all metatarsal bones are very atrophic in characteristic man-

ner, the anterior ends being pointed with entirely atrophic heads. The atrophy is most marked laterally, so that the individual bones are cone-shaped.

Os metatarsi I is 3.5 cm. long, somewhat thin, anteriorly in bony ankylosis with Phalanx I, which is 2.2 cm. long, and in turn, ankylosed to Phalanx II, which measures 1.8 cm. long. Both are pointed anteriorly.

Os metatarsi II is 4.5 cm. long, pointed, and attached anteriorly by a lax-joint-capsule to a thin atrophic Phalanx I scarcely 2 cm. long.

Os metatarsi III is 4.2 cm. long, pointed, articulates with a thin phalanx 2 cm. long, which has a thin, bony shell attached to its head.

Os metatarsi IV is 4.3 cm. long, pointed, without phalanges.

Os metatarsi V is 5.5 cm. long, and has a single phalanx, which is awl-shaped anteriorly.

Specimen 8.—Considerable concentric atrophy of the metatarsal bones and phalanges (Fig. 8). Left foot of a patient with lepra anesthetica beginning in

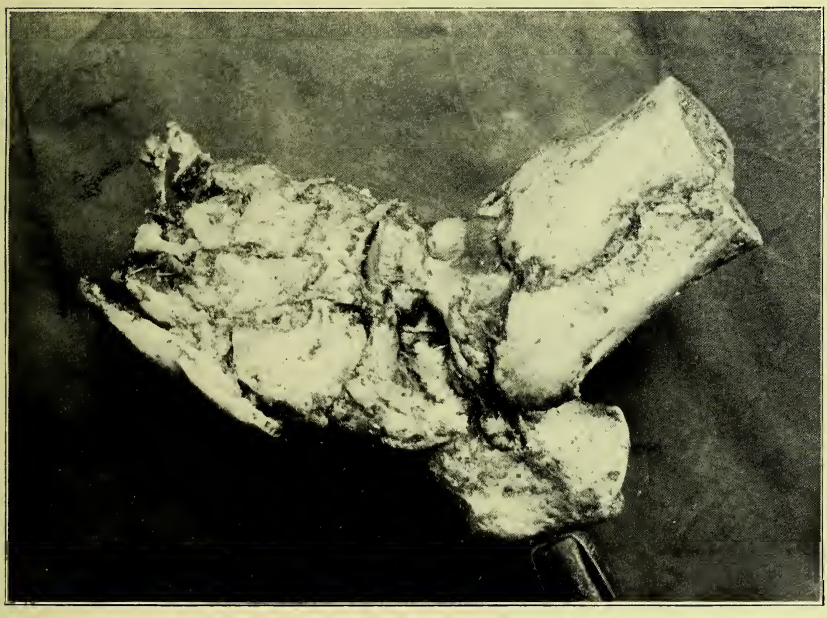

Fig. 12.—Left foot; chronic deforming arthritis of tarsal joints; atrophy of metatarsal bones and phalanges.

1849. On account of ulceration and persistent pain the left foot was amputated in 1888, or thirty-nine years after the onset of the disease. The foot is short and much deformed, chiefly on account of considerable atrophy of the metatarsal bones and, to a lesser extent, of the phalanges. The tarsal bones are normal with the exception of the cuboid, which is short, 1.5 cm. long, and compressed with somewhat rough edges. The metatarsal bones show considerable changes, being in part shortened, narrow and thin, and in part almost entirely resorbed. The same is true of the phalanges, of some of which only rudiments are found. There is no evidence of necrosis or ulceration either in the metatarsal or the phalangeal bones.

Os metatarsi I, 4 cm. long; Phalanx I, 1.7 cm.; Phalanx II, 2 cm. Bones about normal in form and size.

Os metatarsi II, 4 cm. long, thin and pointed; Phalanx I, 2.2 cm.; Phalanx II, 1.2 cm.; Phalanx III, 1 cm. The phalanges about normal; flexed.

Os metatarsi III is 3.5 cm. long, awl-shaped anteriorly with two fragments of phalanges, respectively 1 and 0.8 cm. long.

Ossa metatarsi IV and V have almost disappeared; represented by fibrous cords with small pieces of bones. Phalanges also represented by fragments not over 1 cm. long.

Specimen 9.—Ulcerative necrotic inflammation of the tarsus (Fig. 9). Atrophy of several metatarsal bones. Right foot obtained from a patient with lepra anesthetica with onset about 1877 and amputation of the foot about fifteen years later. The foot is considerably shortened and deformed, chiefly from severe atrophy and destruction of the tarsus, and partly from atrophy of the metatarsal bones and phalanges. Of the tarsus only the posterior part of the calcaneum

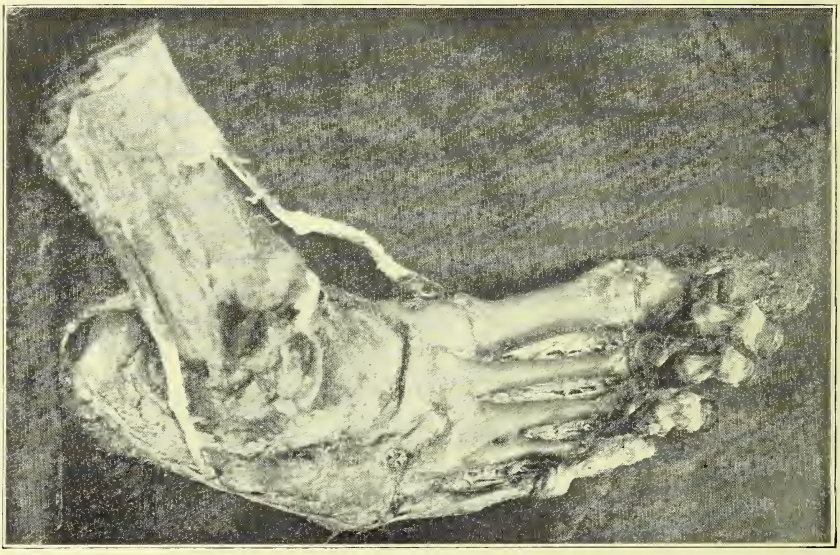

Fig. 13.—Right foot; atrophy of tarsus with arthropathies; concentric atrophy of third, fourth and fifth metatarsal bones.

remains. The talus is much deformed. The os naviculare is very defective and irregular in shape. The cuboid is missing and, of the cuneiform bone, only small spicules remain joined into a plate. Probably ulceration and necrosis of the bones had caused these deformities. The metatarsal bones also show great changes, especially the last three, which are shortened, pointed, and partly fused posteriorly. The phalanges, on the other hand, are mostly normal.

Os metatarsi I, 5.5 cm.; Phalanx I, 2.6 cm.; Phalanx II, 1.8 cm.; normal.

Os metatarsi II, shaft shortened, 5.9 cm.; Phalanx I, 2.1 cm.; Phalanx II. 1.1 cm.; Phalanx III, 0.8 cm.

Os metatarsi III, 5 cm.; Phalanx I, 2 cm.; Phalanx II, 1.1 cm.; Phalanx III, 0.9 cm.

Os metatarsi IV, 3.2 cm.; Phalanx I, 2 cm.; Phalanx II, 1 cm.; Phalanx III, 0.8 cm.

The third and fourth metatarsal bones are fused posteriorly, thin, and greatly shortened. On the outerside, they are joined by a bony plate 1.5 cm. long, which is probably a rudiment of the fifth metatarsal bone. At the anterior end of this bone is attached the first phalanx, 2 cm. long, with the second phalanx 1 cm. long.

Specimen 10.—Lepra mutilans of one foot following an ulcerative inflammation. Also concentric atrophy of the metatarsal bones and phalanges (Figs. 10 and 11). The specimen was obtained from a patient with lepra anesthetica who contracted the disease in 1852 and died in 1888. There had been ulceration of the sole of the left foot. The big toe had been disarticulated in 1882. The left foot was secured at the necropsy. It is greatly deformed, forming a lumpy mass, which is in strong, plantar flexion at the ankle. The tibia and fibula are fused for a distance of 5 cm. at the lower end, forming one pointed bone which has united to the remains of the tarsus. The latter consists mainly of the greater part of the calcaneum (7 by 4 by 3.5 cm.) of entirely irregular shape, and pointed anteriorly. To its inner side there adheres a small, smooth, flat piece of bone,

Fig. 14.—Skiagraph; same as Fig. 1.

evidently remains of the talus, and a splinter of bone adheres on the outside (remains of cuboid?). In addition, there remains of the tarsus only a flattened finger-bone, 3.5 by 2.5 cm., the origin of which can no longer be recognized. These marked changes in the tarsus are probably connected with the ulcerative and inflammatory process in the planta pedis. Of the metatarsus also, only fragments remain, partly in bony connection with the tarsus. The first metatarsal bone is missing, having probably been removed with the hallux. The second metatarsal bone is 2 cm. long and evenly reduced in all dimensions with a relatively large head. The third metatarsal bone is much smaller and thinner, and of the fourth one, only minute pieces are found. The fifth is unrecognizable. The toes are spread out in a fan-shaped manner from the small atrophic tarsus. The hallux is missing. In the second toe the first phalanx measures 2.4 cm. in length and is very thin at its middle; the second phalanx 0.7 cm. long and the third phalanx 0.7 cm. long, are about normal in size and shape. In the third toe, the first phalanx is 3 cm. long, thin and evenly atrophic; the second phalanx 0.9 cm. long, and the third phalanx 1.2 cm. long, are of about normal size and shape.

The first phalanx of the fourth toe is 1.5 cm. long and evenly reduced in all dimensions; the second phalanx is 1.2 cm. long; the third phalanx is missing. The first phalanx of the fifth toe is replaced by a fibrous cord and continues with the second phalanx which is 0.9 cm. long; the third phalanx is missing.

Specimen 11.—Chronic deforming arthritis of the tarsal joints. Typical atrophy of the metatarsal bones and phalanges (Fig. 12). The specimen, obtained from Lepra Hospital No. 1 in Bergen, is the left foot of a patient with lepra anesthetica which commenced in 1862. He died of pneumonia in 1889. There were three ulcers on the left foot and necrosis of the fourth toe. The left foot, which was secured at the necropsy, is greatly deformed and markedly shortened on account of shortening or complete absence of metatarsal bones and phalanges. All of the tarsal bones are intact and of about the usual size and shape. The calcaneum being 7 cm. long, the talus 5 cm. and the cuboid 3.2 cm. They are

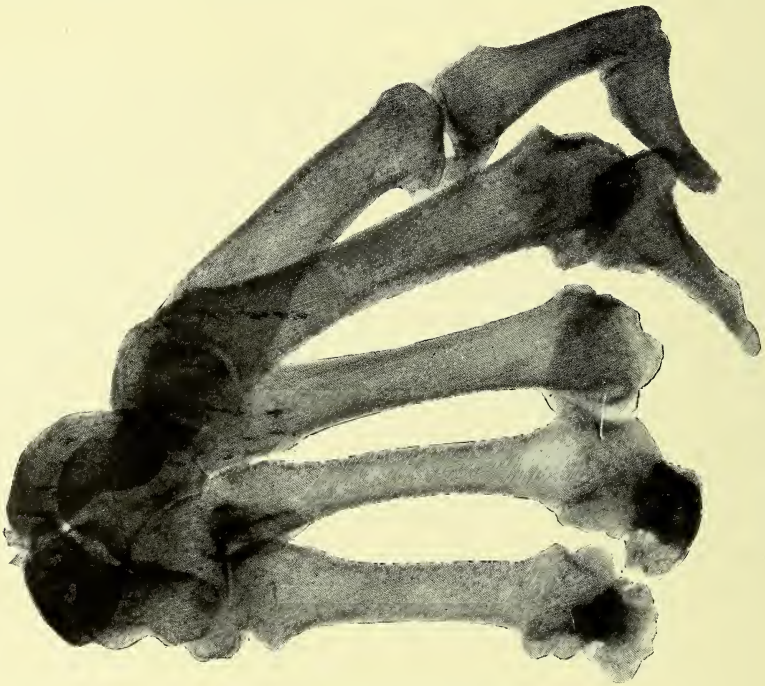

Fig. 15.—Skiagraph; same as Fig. 2.

partly grown together and have rough surfaces which, to some extent, is due to a deposit of osteophytes at the articular ends (chronic deforming arthritis). All of the metatarsal bones are much changed and atrophic, especially at the anterior ends, which are pointed, almost awl-shaped, with their heads missing. The phalanges are also atrophic and pointed anteriorly, especially in the first three toes.

Os metatarsi I, 1.5 cm. long, with one pointed phalanx.

Os metatarsi II, 2 cm. long, with two, thin, pointed, crooked phalanges.

Os metatarsi III, 3 cm. long, with two atrophic phalanges.

Os metatarsi IV, 3.2 cm. long.

Os metatarsi V, 6.2 cm. long, about normal length.

Specimen 12.—Atrophy of the tarsus with joint affections. Typical concentric atrophy of third, fourth and fifth metatarsal bones (Fig. 13). The right foot with joints and tendons from a case of leprosy of the smooth variety. The foot is a good example of the joint affection so frequently encountered in leprosy, and also presents some atrophy of the metatarsal bones. The ankle-joint is the seat of severe arthritis with marked dilatation of the joint cavity and proliferation of the synovial membrane. The tarsal bones show marked changes, consisting both in atrophy and proliferation of bone, particularly in the vicinity of the

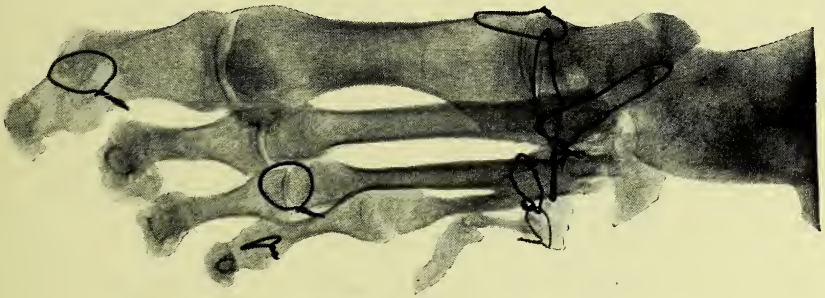

Fig. 16.—Skiagraph; same as Fig. 5.

joint. The talus is deformed, its body particularly being atrophic and reduced to a flat, bony plate. For this reason the lower end of the tibia has pressed into the tarsus. Of the os navicularis, only narrow rims externally and internally remain, which are united by a lamellar central portion. The cuneiform bones are barely demonstrable, merely formed by irregular pieces of bone fused with the first three metatarsal bones. The cuboid is atrophic, deformed, and entirely

Fig. 17.—Skiagraph; same as Fig. 6.

fused with the calcaneum. On account of these severe changes in the tarsus the foot has sunken down and is in pronounced valgus position. The metatarsal bones and phalanges are somewhat atrophic, especially those of the fourth and fifth toes.

Os metatarsi I, 6 cm. long; Phalanx I, 2.8 cm.; Phalanx II, 2.5 cm.

Os metatarsi II, 6.5 cm.; Phalanx I, 3 cm.; Phalanx II, 1.8 cm.; Phalanx III, 1.4 cm.

Os metatarsi III, 5.8 cm.; Phalanx I, 2.4 cm.; Phalanx II, 1.6 cm.; Phalanx III, 1.6 cm.

Os metatarsi IV, 5.3 cm.; Phalanx I, 2.8 cm.; Phalanx II, 1 cm.; Phalanx III, 1.4 cm.

Os metatarsi V, 5.5 cm.; Phalanx I, 2 cm.; last two phalanges rudimentary.

The third and fourth metatarsal bones are fused at their base, somewhat pointed anteriorly, and flattened from side to side. Seen from above they appear as sharp edges. Also the fifth metatarsal bone is thin, and atrophic anteriorly. The phalanges on the whole are fairly normal.

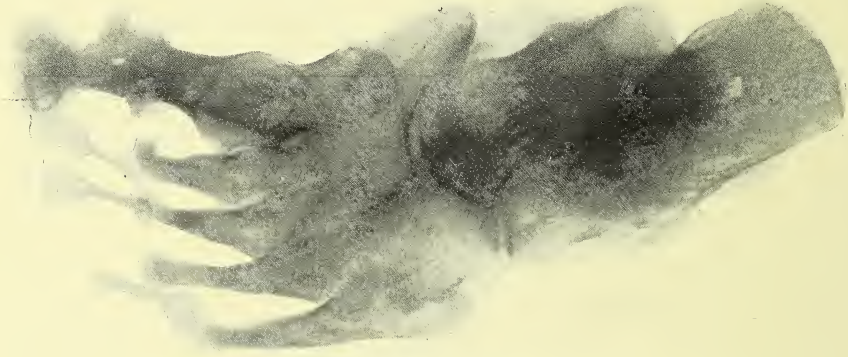

Fig. 18.—Skiagraph; same as Fig. 7.

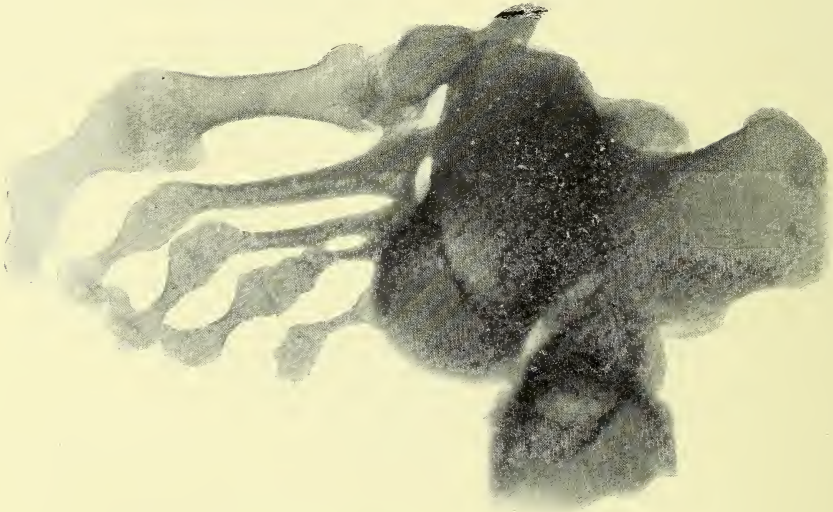

Fig. 19.—Skiagraph; same as Fig. 8.

I shall first give a collective description of the osseous changes although the illustrations convey the best idea of them.

There is an atrophy of the bones of the hands and feet which may proceed to such a degree that certain bones finally entirely disappear.

The atrophy principally affects the most distal bones, the phalanges, metacarpals, and metatarsals; it is little pronounced in the tarsus and carpus. The bones may atrophy in their entirety, though sometimes more so in the shaft, and become thin and brittle, but their shape is preserved. This variety resembles the so-called atrophy of disuse, though the bones generally are somewhat decreased in size. In the more characteristic form the atrophy is concentric and chiefly involves the most peripheral parts of the phalanges, metacarpal, and metatarsal bones. These bones become considerably shorter than normal, taper anteriorly in a cone-like manner, and may become awl-shaped; or the atrophy is most pronounced on the sides, so that the bones become plate-like with sharp edges above and below. The distal heads of the metacarpal and metatarsal bones usually

Fig. 20.—Cast of left hand; lepra mutilans.

disappear, while at their base the bones preserve their usual width and thickness. The surface is generally smooth. The shortening of the metacarpal and metatarsal bones may be very considerable and most pronounced either in those on the outside or those on the inside. It may be so marked that the length is only half the normal one, or even less (Specimen 6). The phalanges atrophy in a similar manner, become very thin, pointed anteriorly, and porous. Finally only disconnected remnants are left which ultimately may be entirely absorbed so that the toes become shapeless lumps without any bony framework. Our specimens illustrate this condition very well.

In the joints, various changes are also seen, on the whole resembling those of arthritis deformans. Especially in the joints of the tarsus,

we see a form of chronic inflammation of the joint-capsules with the picture of pronounced, proliferating synovitis and serous effusion (Specimen 12); at the same time, the articular ends show changes, such as atrophy of cartilage and bone, or proliferative alterations in the form of nodular projections, osteophytes, etc. The osseous substance, as a whole, in most cases is the seat of marked osteoporosis. The tarsal bones become brittle and often are squeezed together to form thin plates of bone. If the tarsus is the principal seat of atrophy, the whole foot may sink down in a pronounced valgus position. Similar signs of chronic joint disease are seen in atrophic, leprous hands, but less frequently and less prominently.

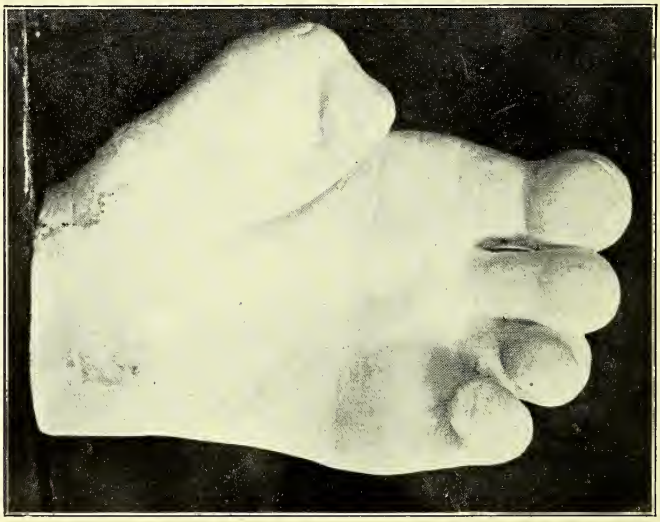

Fig. 21.—Cast of left hand; lepra mutilans.

The question arises whether this peculiar bone atrophy is specific, and directly due to leprous neuritis, or if it can be satisfactorily explained in some other manner.

In the first place it must be emphasized that these atrophies are not due to external inflammation. In other words, they do not belong to the ordinary forms of lepra mutilans dependent on ulceration ("mal perforant") and chronic suppuration, extending to the bones and joints, accompanied by secondary destruction of bone. As shown by the specimens, it is true that in addition to typical atrophy of bone we may also find forms of chronic inflammation which lead to extensive destruction (Specimens 4, 9, and 10), but the inflammation and the necrosis of bone have no

peculiar characteristics and may easily be recognized. They are also found in localities where there is no atrophy, and may be found wanting in the presence of typical concentric atrophy. This mode of explanation must, therefore, be excluded.

Nor can we be dealing with pure atrophy of disuse, as we then would expect to find all the bones of the foot atrophic and, what is most important, the atrophy in individual bones ought to be evenly distributed, not limited to certain parts. One would expect an anatomic picture similar to that seen in paralyzed limbs, for instance, after an attack of acute poliomyelitis. But this is not the case. In paralyzed limbs the bones usually retain their shape and ordinary size, but merely become brittle and osteoporotic, while circumscribed atrophy such as here described is

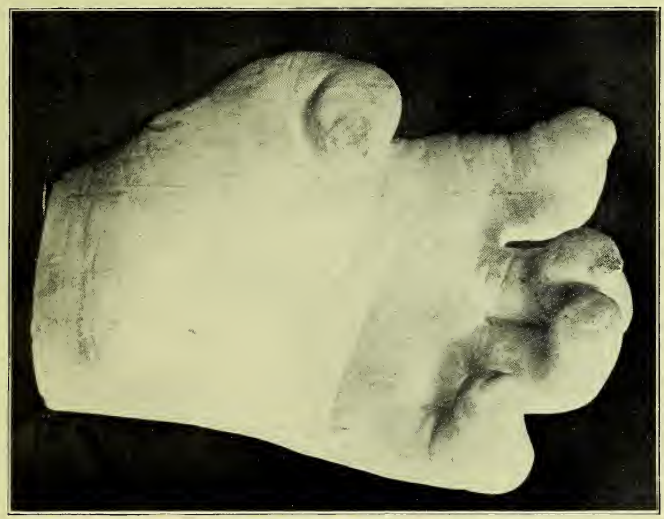

Fig. 22.—Cast of left hand; lepra mutilans.

entirely absent. It is another matter that disuse may play a considerable rôle in the development of atrophy.

Finally, it might be maintained that the atrophy might depend on genuine, specific, leprous osteomyelitis and periostitis. Lepra bacilli and leprous granulation-tissue in the osseous system have been demonstrated chiefly by Hirschberg and Bichler[3] and, in that case, it might be supposed that absorption of bone might result in atrophy. *A priori,* the objection could me made that in such a case concentric atrophy would be less likely to occur than the destruction of bone similar to that seen in tuberculosis

3. Hirschberg and Bichler: Lepra der Knochen, Dermat. Ztschr., 1909.

and syphilis. But skiagraphs taken of all of our cases show that there is no trace of such central lepromata or of any periostitis. This has already been pointed out by Deycke,[4] who also studied skiagraphs of leprous bones. He also could demonstrate a peripheral atrophy without reactive changes, but he had no opportunity to make anatomic studies of the skeletons of the hands and feet, or to make histologic examinations.

The microscopic studies made to clear up this question, which were especially thorough in the case of Specimen 12, argue against the position that we are dealing with results of leprous inflammation. Thus in speci-

Fig. 23.—Cast of right hand; lepra mutilans.

mens of atrophic toes, we find small pieces of bone with uneven surfaces containing osteoclasts in the depressions, but no leprous osteomyelitis or periostitis could be demonstrated and no lepra bacilli were found.

Therefore, no weight can be given to any of these objections. There is no good reason for doubting that this bone atrophy is really of neurotrophic origin, and everything seems to point to leprous interstitial neuritis as the cause. As has been strongly contended by H. P. Lie,[5] it

4. Deycke: Knochenveränderungen bei Lepra nervorum in Roentgenograph, Fortschr. a. de Geb. d. Roentgenstrahlen, ix.

5. Lie, H. P.: Lepra im Rückenmark und den peripheren Nerven., Vienna and Leipsic, Wilhelm Braumüller, 1904.

must be maintained that the trophoneurotic changes are associated with advanced leprous neuritis of the mixed nerves, and not with a definite form of the disease, or with particular lesions of the central nervous system, or certain nerves.

In this connection, it must be remembered that similar concentric atrophy of the toes has been described as following nerve lesions of various kinds, such as gunshot wounds, causing destruction of peripheral nerves, in which case the atrophy was characterized by the toes being shorter and more pointed at the ends just as we see in characteristic leprous bone atrophy.

Fig. 24.—Cast of right foot; lepra mutilans.

We must also consider the cutaneous changes occurring in leprous fingers and toes. The skin becomes thin, atrophic, smooth and glistening ("glossy skin"), with atrophy of epidermis, cutis and subcutaneous tissues and with disappearance of glands and loss of hair. It has been noted in our cases that the skin finally has become scar-like and inelastic. The nails also undergo changes as they cease to grow and fall off, or become corrugated, rough, and chipped, or greatly thickened and nodular. It may be generally known, as has been shown by Hansen and Looft,[6] that these cutaneous changes are due to leprous neuritis and are trophо-

6. Hansen and Looft: Die Lepra vom klinischen und pathologisch-anatomischen Standpunkte, Biblioth. med. int., 1894, ii.

neurotic in nature and comparable to the concentric atrophy of bones. As is well known, analogous changes in the skin may follow lesions of peripheral nerves.

The question arises whether the explanation of this characteristic atrophy of bone should be looked for in special trophic nerves. Aside from the fact that these nerves are still entirely hypothetical, at least not proved experimentally, it must be sufficient to recall the influence exerted by all forms of nerves, motor, sensory and vasomotor, on tissues and organs. As the leprous neuritis involves all nerves without selection, it must be supposed that all forms of nerve fibers suffer from the chronic inflammation and that this will express itself in loss or alteration of the influence normally exerted by the nervous system on nutrition. This must be considered sufficient explanation; in other words, the leprous.

Fig. 25.—Cast of left foot; lepra mutilans.

trophoneurotic bone-atrophy does not require the assumption of the existence of trophic nerves.[7]

When we study histologic specimens of chronic interstitial leprous neuritis and note the marked alterations in the nerves, we readily appreciate the influence on the tissues exerted by such chronic inflammation with progressive degeneration and atrophy of nerve fibers going on for many years, and will not hesitate to look upon atrophy of the osseous system as a result.

As the opportunities for anatomic study of leprous neuritis is not as good elsewhere as in Norway, where there still is considerable, though

7. In this connection the reader is again referred to my previously mentioned work, Om. path. an forandringer af neurotrofisk oprindelse, 1900.

rapidly decreasing leprosy, I shall briefly relate a recent case of lepra mutilans with marked deformities, especially of the hands.

The case was that of a woman 72 years old, who was treated for leprosy during the last twenty-one years of her life in the dermatologic ward of the government hospital in Christiania. The onset was a few years before admittance to the hospital with anesthetic area and later cutaneous nodules on the back, face and particularly the dorsum of the hands and forearms. Gradually lepra mutilans with considerable deformity of the hands developed. Before death marasmus had set in and the patient also had developed leprous inflammation of the larynx, pharynx, and mouth. The necropsy revealed changes in all the nerves examined of the right forearm and hand, especially thickening of the nerves, which increased towards the wrist in the case of the radial and median nerves

Fig. 26.—Left hand; lepra mutilans.

but which was most marked at the elbow in the case of the ulnar nerve. The nerves were also firm and fibrous, and, in the case of the distal part of the radial nerve, adherent to the surrounding tissues, and some of them, especially the radial nerve, contained brown or yellowish-brown deposits ("brown bodies," i. e., large heaps of bacilli) The histologic findings are as follows:

The median nerve near the wrist showed concentric thickening of the perineurium of each nerve bundle and slight lymphocytic infiltration; also increased interstitial tissue within the bundle, i. e., endoneuritis. There was marked thickening of the endoneurium in which, only here and there, few and partly atrophic nerve fibers were seen. There was marked round-cell infiltration of the ulnar nerve at the wrist. The interstitial tissue was increased and almost no

nerve fibers were demonstrable. Scattered in this granulation-tissue, there were numerous heaps of bacilli which stained red and were generally intracellular. In the radial nerve near the wrist there was proliferating connective tissue without nerve fibers with scanty lymphocytic infiltration, and exceedingly numerous, and large heaps of bacilli generally arranged longitudinally and located intracellularly.

May the articular changes described also be looked on as genuine trophoneurotic affections? This is considerably more doubtful. On account of their whole appearance they are analogous to the arthropathies accompanying certain diseases of the brain and cord, such as tabes dorsalis and syringomyelia. It is especially to the credit of Charcot to have called attention to them and to have pointed out that the arthropathies

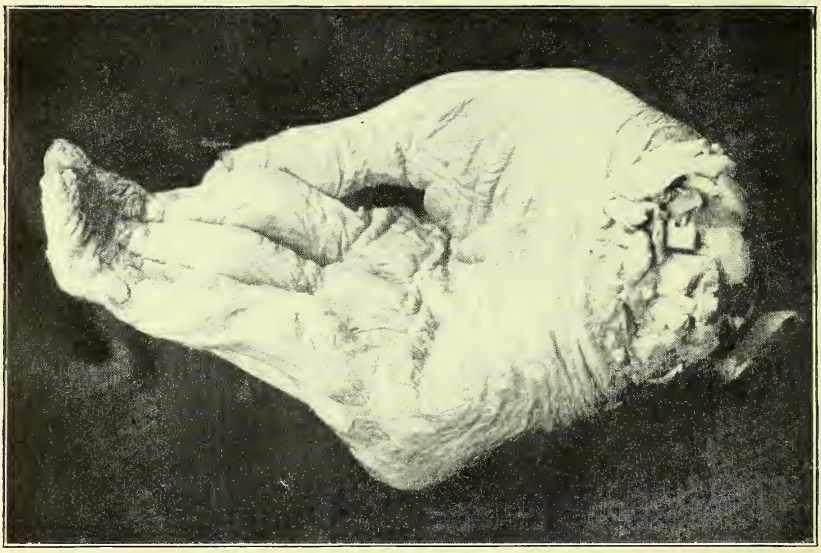

Fig. 27.—Right hand; lepra mutilans.

are specific, trophic phenomena dependent on a direct nervous influence exerted by disease of the central nervous system, particularly the anterior grey horns of the cord. H. Heiberg also seems to have shared this opinion.

The findings, however, particularly those in the cord, on which Charcot and others based their hypothesis, have not been confirmed by other observers. For this reason the condition of the peripheral nerves has been looked to for explanation, and in the case of leprosy the leprous neuritis.

The question arises whether it is not more natural and correct to consider these arthropathies, and particularly the joint affections of

leprosy, as merely indirectly caused by the nervous disease. In the case of the leprous arthropathies, we must bear in mind the considerable degree of muscular atrophy produced by the neuritis which renders the joints more lax and the movements ataxic. The development of arthropathies is further favored by the condition of the bones which become brittle, osteoporotic, and deformed and easily give way to pressure and trauma of any kind. The existing analgesia and anesthesia are also of importance. The sensory nerves supplying the joint-capsules and ends of the bones are affected so that the ordinary sensation produced by the abnormal position of the joints is lost, which predisposes to ataxic movements and anatomic changes. The reflex influence of the centripetal nerves on the normal nutrition of the tissues is lost, and this also exerts its influence.

All these factors are of importance and by their indirect influence on the joints may result in the articular affections described, without it being necessary to look for the explanation in a special trophoneurotic influence.

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